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Lupus pernio: a tale of four characters in search of a malady

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Introduction

Lupus pernio is the most typical and easily recognizable skin lesion of sarcoidosis. Ernest Besnier in 1889 gave the lesion its unusual name and distinguished it from the lesion of tuberculosis, called lupus vulgaris.

The colorful history of the lesion is a human tale involving dedicated physicians and their grateful patients.

Comment

Ernest Besnier (1831-1909)

Honfleur, an old picturesque port, sits on the southern estuary of the Seine, one of the principal rivers of France. Many artists, including Gustave Courbet, Claude Monet, John Jongkind and Eugene Boudin have immortalized its beauty, serenity, and natural elegance. The explorer Binot Paulmierde Gonneville set forth from its shores in 1503 to explore Brazil. Another navigator, Jean Denis left Honfleur in 1608 to found Newfoundland and the mouth of Saint Lawrence River. Other famous Honfleurais have included Alphonse Allais, writer and humorist: Erik Satie, musician; and Stephane Ferrand, wildlife photographer. In 1859, Baudelaire ravaged by his illnesses, long-term use of laudanum and absinthe, stress and poverty, moved to Honfleur to live with is mother. He was at peace in the seaside town where wrote *Le Voyage*.

Ernest Besnier, one of the characters of our tale was also born in Honfleur on April 21, 1831 (Fig. 1). His father was a government official on the move. As a result, the young Besnier was schooled in different places, including Honfleur, Marseilles, and Orleans. Finally, he settled in Paris to study Medicine. He worked hard and won the first prize of the Internat des Hopitaux de Paris in 1853. He was then 22 years old. In 1867, at 32 years of age, Besnier secured his Doctor of Medicine degree and joined the Institution Saint Perrine. He stayed there for 10 years and published several epidemiological reports on the then prevalent diseases. In 1873, he was moved to the Hopital Saint Louis to occupy the Chair in Dermatology vacated by the retirement of Bazin.

An intellectually honest man, Besnier worked for a quarter of a century in complete harmony with his medical colleagues and his patients. He was an outstanding teacher, had a large clinical practice, and was an able administrator and organizer. He built an annex to the hospital with complete histopathology and parasitology laboratories. He introduced the word "biopsy" to describe the tech-

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Fig. 1. Ernest Besnier

nique of taking skin samples in order to study histopathology. He founded the medical journal "Annales de dermatologie et de syphilographie" with Pierre Adolphe Adrien Doyen and translated Moritz Kaposi's famous book on skin diseases from German to French.

On February 14, 1889, Ernest Besnier, now Chief of Medicine and one of the grand men of French Dermatology presented a case entitled, *"Lupus Pernio of the Face- symmetrical scrofulo-tuberculosis of the upper extremities"*. The lecture began, "Here is a man, aged 34 years, who exhibits lesions of the same type in both face and upper extremities, although with differing appearances in each, which are not well understood or described. First the face represents Lupus erythematosus, which I propose to name Lupus Pernio or Lupus asphyxiation, similar to Hutchinson's chilblains, but not identical."

In describing lupus pernio, Besnier clearly differentiated the lesion from other dermatological conditions, including tuberculosis. Ernest Besnier died in Paris on May 15, 1909 leaving a simple epitaph, "For thirty years he shed an incomparable luster on the teaching at the Saint Louis Hospital" (1).

Lucien Marie Pautrier (1876-1959)

Pautrier was born in Marseille on August 2, 1876. He did his early schooling and first year of medical education at his birthplace, but moved to Paris to complete his medical training. He became interested in dermatology.

In 1903, he defended his 350-page thesis, "Atypical Tuberculosis of the Skin" in which he classified various skin manifestations of tuberculosis. The work was so important that he was allowed to continue clinical research under Louis-Anne Jean Brocq (1856-1928) at L'Hopital Saint-Louis. During the First World War, Pautrier was mobilized in an artillery regiment where his bravery won him a citation of the Order of the Armed Forces and the Cross Chevalier of the Legion d'Honneur. He became the Chairman of Clinical Dermatology at the University of Strasbourg in 1919. In 1939, because of the War, Pautrier had to flee to Dordogne and then to Switzerland where he stayed till the end of the Second World War.

In 1946, at age of 70 years he returned to Strasbourg and resumed the Chairmanship.

Pautrier's research was admired for its vitality, clarity, and elegance of style. He refuted the role of the tubercle bacillus in the pathogenesis of lupus erythematosus. He was the first to oppose the idea that tubercle bacillus was the cause of lupus pernio and sarcoidosis. He linked lupus pernio with sarcoidosis and presented sarcoidosis as a complex illness caused by one agent. He was the first to describe the reticulo-nodular feature of pulmonary sarcoidosis and showed that parotid involvement and uveitis were expressions of sarcoidosis.

Tenneson first described the histology of lupus pernio, but it was Pautrier who deduced the systemic nature of the disease from studying the histopathology of lupus pernio and other skin lesions. He continued the outstanding tradition of sarcoidosis research, initiated by Besnier. In 1939, he published a book, "The disease of Besnier, Boeck-Schaumann". His publications used wax models depicting lupus pernio and other skin lesions (Fig. 2).

Pautrier was highly respected by his colleagues. His life embodied wisdom, enthusiasm, generosity, and passion for social justice. He is buried in Marseille, on Provencal soil, as was his desire.

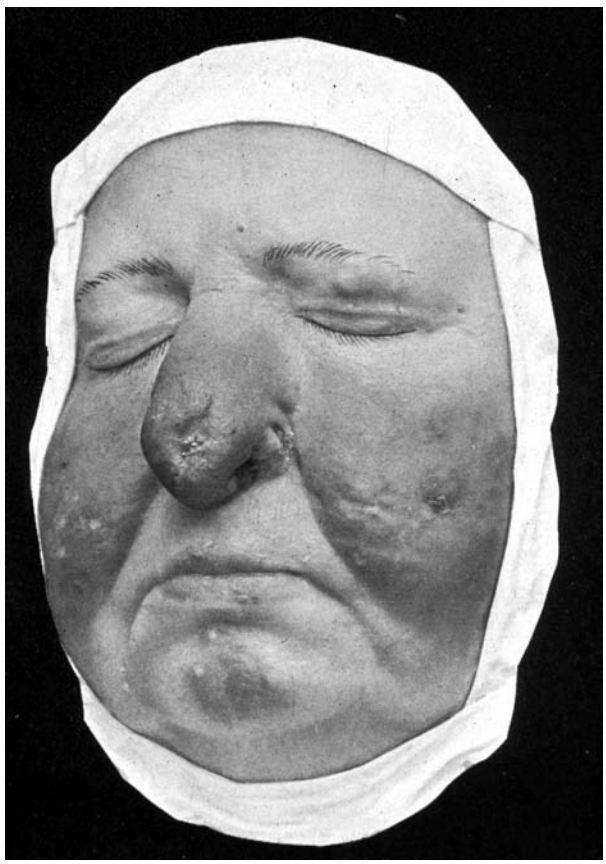

Fig. 2. Lupus pernio, wax model

Mrs. Mortimer

In 1898, Jonathan Hutchinson used the term "Mortimer's malady" to describe the disease that had afflicted Mrs. Mortimer, a 65-year old Londoner. Hutchinson observed that the skin lesions had been present for a year and consisted of patches on her cheeks and on the back of the upper arms. Although the lesions were symmetrical, they were larger and more abundant on the left cheek and right arm rather than on the opposite parts. They were dusky red in color, well defined, raised and soft (Fig. 3). Six months later these patches had increased in number and size. The lobule of the right ear was affected, and the nose was much swollen across the bridge. There was slight scaling but no ulceration, pustulation or scarring.

At this stage, Hutchinson demonstrated the case to the Dermatological Society of London where it was urged that portions of her skin should be removed for microscopic examination. Hutchinson recorded

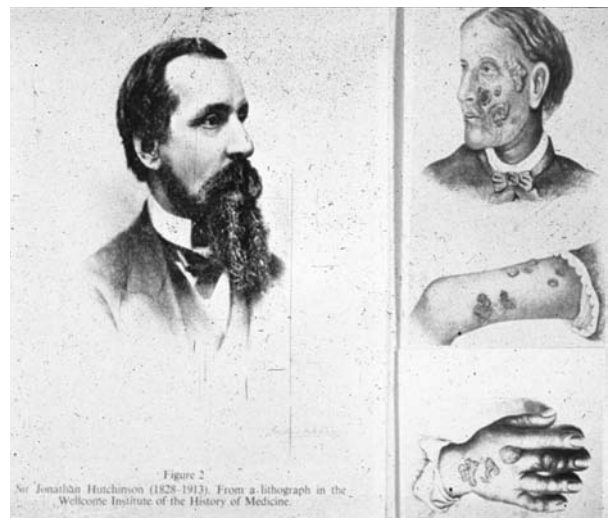

Fig. 3. Mrs Mortimer`s lesions

that he subsequently suggested the plan to the patient, with the result that he did not see her again. Thus Hutchinson was deprived of priority in the histological description of what most likely was the first case of lupus pernio. Hutchinson thought that *Mortimer's malady* may not improbably be a tuberculous affection and one of the lupus family, but if so it differed widely from all other forms of lupus, both in its features and course'. He called it 'lupus vulgaris multiplex non-ulcerans et non-serpiginous' (Fig. 4). Hutchinson did not use the term lupus pernio (2, 3).

Mrs. Amy Verrant

Mrs Amy Verrant, a true Cockney, was born in 1900 in the East End of London within earshot of the Bow Bells, i.e. the bells of St. Mary-le-Bow church in Cheapside in the City of London. She had a healthy carefree childhood before the First World War. In the nineteen twenties young Amy developed redness of her eyes,blurring of vision, and weakness in her face. Slowly, she became blind, developed skin lesions and swellings of her fingers.

The specialist medical care was not easily available during and many years after the World War II. Bow bells were silenced. Amy Verrant endured her illness in silence. In the late nineteen fifties, Mrs Verrant showed up at the Royal Northern Hospital Sarcoidosis Clinic which was run by Dr. David Geraint James, an ebullient Welsh physician who later be-

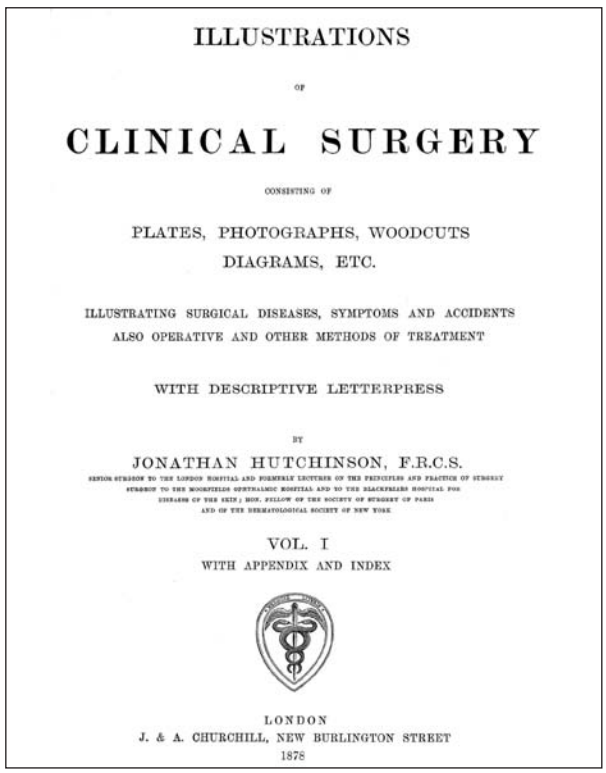
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Fig. 4. Hutchinson's Illustrations of Clinical Surgery

Fig. 5. Amy Verrant

came founder and President of the World Association of Sarcoidosis and Other Granulomatous Disorders (WASOG) and Editor of Sarcoidosis Vasculitis and Diffuse Lung Diseases. Dr. James spread the gospel of sarcoidosis to all corners of the world. His most dramatic picture-slide depicting the course of chronic sarcoidosis was that of his patient Amy Verrant. The picture displayed impressive lupus pernio, blindness, facial palsy, and sausage-shaped fingers (Fig. 5). She also had mild pulmonary fibrosis.

Mrs Verrant became a household word in the international sarcoidosis family. She regularly attended James' Royal Northern Hospital clinic and met many of the local and international sarcoidosis specialists who visited the clinic. She regaled them with stories of the chronic infirmity laced with her charming Cockney humor. Mrs Verrant took great interest in the disease and was, arguably, the first patient-advocate for the disease. The graceful and generous woman died at the age of 90-years. James established Amy Verrant Sarcoidosis Research Fund and used it to support young sarcoidosis researchers working at the Royal Northern Hospital Clinic. The Hospital, the Clinic, Amy Verrant and the Amy Verrant fund are now part of history.

Lupus pernio

Lupus pernio, the most characteristic of all sarcoidosis skin lesions, is a chronic, violaceous, indurated skin affliction with a predilection for the nose, ears, lips, and face. In the United States of America the lesion is more frequent in African American women than in Caucasian patients. In the United Kingdom, lupus pernio is more common in women of West Indian origin.

The appearance of lupus pernio ranges from a few small button-like nodules under the tip of the nose to exuberant plaques covering the nose and spreading across the cheeks. There may be similar nodules on the eyelids and ears and associated plaques on the arms, buttocks and thighs. It may be complicated by nasal ulceration and septa perforation. The lesion is associated with chronic fibrotic disease in many systems.

James reported thirty-five patients with lupus pernio in a series of 818 patients seen at the Sarcoidosis Clinic at Royal Northern Hospital, Lon-

don. The analysis provided the natural history of the affliction and associated clinical and radiological features. Lungs were involved in 74% of patients; upper respiratory tract in 54%, reticulo-endothelial system in 54%, bone cysts in 43% and ocular lesions in 37% (4). Lupus pernio is a convenient experimental model to assess clinical course and response of therapy in sarcoidosis. In a single patient one can study the morphologic appearance of the lesion, histological features of the nasal mucosa, and radiographic changes of the nasal bone erosion (5).

The treatment of lupus pernio needs to be prolonged, aggressive, and under continuous supervision. The drugs commonly used are corticosteroids, methotrexate, hydroxychloroquine, infliximab, and thalidomide. Cosmetic camouflage improves the quality of life and social comfort (6).

The end

The story of lupus pernio is an integral part of the history of multisystem sarcoidosis. It is easily recognizable and; it can be visualized and measured. No specific and effective treatment is available for lupus pernio, but the lesion remains an experimental model for future clinical studies.

References

1. Turiaf J. Ernest Besnier (1831-1909) Et Lucien-Marie Pautrier (1876-1959). In Levinsly L, Macholda F (Eds) Fifth International Conference on Sarcoidosis. University of Karlova, Prague 1971; 57-62.
2. Hutchinson H. Jonathan Hutchinson: Life and Letters. WM Heinemann Medical Books Ltd. London, UK 1946.
3. Hutchinson J. Case of Livid papillary psoriasis. Illustrations of Clinical Surgery. J & A Churchill Ltd, London 1877; 1: 42.
4. James DG. Sarcoidosis and Other Granulomatous Disorders. Major Problems in Internal Medicine. W.B. Saunders Co. Philadelphia, 1985: 24.
5. James DG. Lupus pernio is a valuable experimental model. Sarcoidosis: Ninth International Conference on Sarcoidosis and Other Granulomatous Disorders. Pergamon Press, Paris 1981; 465-9.
6. Baughman R, Lower E. Evidence-based therapy for cutaneous sarcoidosis. Clinics in Dermatology 2007; 25: 334-40.